CLASSICAL PRIMARY CHONDROSARCOMA OF THE FALX : CASE REPORT

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RÉSUMÉ : Les chondrosarcomes sont des tumeurs malignes primitives cartilagineuses qui surviennent au niveau des os long et du bassin. Environ 7% de ces tumeurs surviennent au niveau de la tête et du cou. Les chondrosarcomes intracrâniens représentent 0, 16% des tumeurs intracrâniennes et sont généralement localisés au niveau de la base du crâne. Dans de très rares cas ils peuvent avoir une origine méningée : faux, tente ou convexité. Il existe 3 variantes histologiques qui sont le type classique, le type mésenchymateux et le type myxoïde. Nous rapportons le cas d'un patient présentant un chondrosarcomeclassique primitif de la faux traité par chirurgie et qui est vivant sans récidive à 92 mois avec discussion de la littérature.

***Mots clés** : Chondrosarcome, Faux, Chirurgie, Type classique.*

ABSTRACT : Chondrosarcomas are primary malignant cartilaginous tumors that occur in most of cases in epiphysis of long bones and pelvic bones. Near 7% of all chondrosarcomas occur in the craniocervical region. Intracranial chondrosarcomas represent 0.16% of all intracranial tumors and are generally localized at skull base. In very rare instances they can arise from the meninges at the falx, tentorium or cerebral convexity. These tumors have 3 histological variants: classical, mesenchymal, and myxoid. We report a case of classical primary chondrosarcoma of the falxtreated by surgerywho is alive at 92 months without recurrence and discuss relevant literature.

***Key words :** Chondrosarcoma, Falx, Surgery, Classical type*

INTRODUCTION

Chondrosarcomas are mainly bone tumor and only 7% can involve the head and the neck [21]. They represent near 0.16% of intracranial tumors [10] and 75% of them are located at the skull base [12]. In rare instances they can be intracranial extraskeletal arising from meninges and can be misdiagnosed with other extra axial tumors such as meningiomas [10, 16]. There are 3 histological types: classical, mesenchymal and myxoid [4]. Surgery is the mainstay treatment and can be followed in some instances by radiotherapy and chemotherapy [4, 10, 13, 16, 19, 1]. Prognosis factor are extension of resection and histology [1, 4, 12-14, 16, 19, 21]. We report a case of classical chondrosarcoma arising from the falx who has been treated by surgery alone and alive with no recurrence at 92 months.

CASE REPORT

A 33 year-old man was referred to our department for intracranial hypertension on August 2010. He had a one year history of headaches. He was also treated for manic-depressive psychosis. On examination, he was dizziness and there was no motor deficit. Ophthalmologic examination done before admission revealed anomalies of visual field and papillary edema at fundoscopy. CT scan showed a heterogeneous high density mass lesion in right parieto-occipital region with calcifications and cystic areas (Fig. 1).

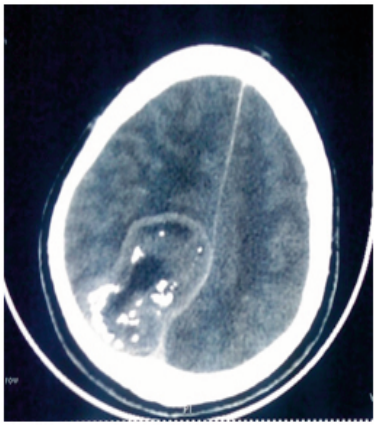

Fig. 1 : Axial CT Scan showing calcifications inside the lesion

Contrast injection revealed heterogeneous enhancement and peritumoral edema was slight. MRI showed a voluminous and heterogeneous lesion attached to the posterior part of the falx with a “honeycomb” enhancement pattern after injection of contrast (Fig.2). Angio MRI revealed an avascular lesion. Radiologist concluded to the diagnosis of meningioma of the falx. The patient underwent a right parieto-occipital craniotomy. There was a well demarcated plan between tumor and parenchyma. The tumor appeared white and lobulated, and had cartilaginous consistence andwe have found inside the lesion a cystic area filled by old blood (Fig. 3). The lesion was inserted on the falx. We have achieved a gross total resection. Histopathology concluded to a classical chondrosarcoma grade II (Fig. 3). Postoperative period was uneventful with recovery of visual field anomalies. We have performed a bone scan to exclude bone localization ; it was normal. Regular follow-up and MRI have been performed. Patient is alive at 92 months with no recurrence on imaging (Fig. 4).

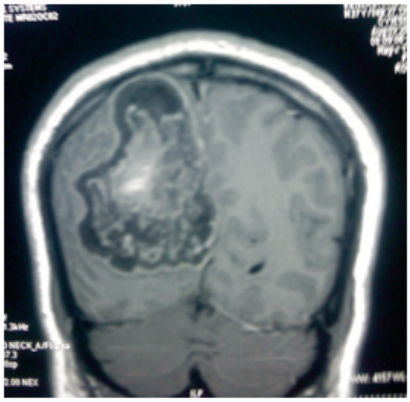

Fig. 2 : MRI T1 Gado coronal view showing “honeycomb” enhancement

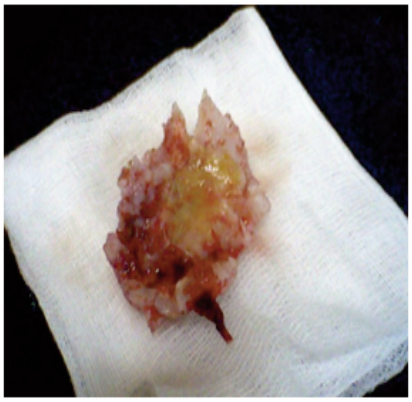

Fig. 3 : macroscopic specimen, the yellow part correspond to the cystic portion

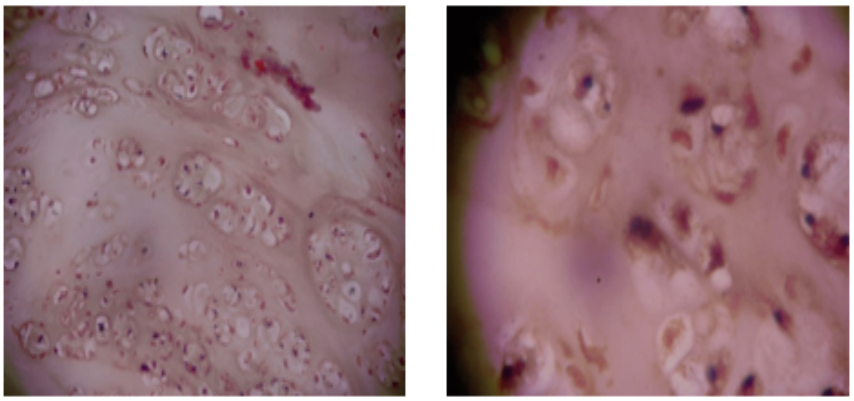

Fig. 4 : microscopic view with HE coloration. A: cartilaginous lobules, high cellular density and calcifications. B: cellular atypia reperesented by bi nucleation

DISCUSSION

Chondrosarcomas are mainly bone lesions. Only 7% of chondrosarcomas involve the head and the neck [21]. Intracranial chondrosarcomas are rare and represent 0.16% of all intracranial tumors [4, 19]. The first description of intracranial chondrosarcoma was made by Mott in 1899 [10]. More than 75% of intracranial chondrosarcomas are located at the skull base [12, 19]. In rare instances, they can be extraskeletal and localized along the falx, tentorium or dural convexity and very rarely be purely intraparenchymal [4, 12, 21]. Case arising from the plexus choroid has also been described [17]. Origin of skull base

chondrosarcomas is from rests of chondrocytes at the synchondroses of skull bones [19] while it is thought that extraskeletal chondrosarcomas arise from pluripotent mesenchymal cells [12] or cartilage rests in the dura [4]. Very rare cases are purely intraparenchymal and are believed to originate from the pia-arachnoid cells of the Virchow-Robin spaces [19].

Intracranial extraskeletal chondrosarcomas are generally supratentorial and a great part of them involve the falx or its neigh borhood [12, 19]. Our patient was a male but a female predominance is described [10, 19]. Most of patients reported are adults between the 3rd and 5th decade but some cases were described in adolescents [5, 9, 14, 22]. On CT scans lesions are isodense to hyperdense including calcifications and heterogeneous enhancement [10]. We have also observed calcifications in our patient. On imaging, intracranial extra skeletal chondrosarcomas are often mistaken as extra axial tumors such as meningioma, hemangio pericytoma, chordoma, solitary fibrous tumor and meningeal metastatic carcinoma [10, 16]. But main differential diagnosis is meningioma [13]. A brighter signal on T2-weighted images, absence of dural tail, minimal peritumoral oedema, and avascularity can make the difference with meningioma but this is not pathognomonic [12]. The presence of “honeycomb” enhancement is another characteristic feature of classic chondrosarcomas [12, 16] this was observes in our patient. Mainstay of treatment is surgery and first choice should be radical resection because prognosis is linked to the extent of the surgical removal [4, 10, 12-14, 16, 19, 21] particularly in classical types as our case. From a histopathological point of view, intra cranial chondrosarcomas are divided in 3 variants: classic, mesenchymal, and myxoid [4]. These variants have different clinical presentation and prognosis [16]. Classical variant has 3 histologic grades : grade I, II, and III ; grade I is similar to benign cartilaginous tumors while grade II and III are more cellular with more mitoses and less chondroid matrix[13]. Published cases of pure falcine chondrosarcomas are resumed in Table 1. At our knowledge there are only 22 cases described before our case. The most frequent histological subtype was the classical which was observed in 12 cases (54.5%). 8 patients have a mesenchymal type and only 2 myxoid case had been reported [1, 21]. Our patient had also a classical type grade II; since it presents cellular atypia. Histology is also a prognosis factor since mesenchymal and myxoid types are more aggressive than classical variant. These types have a tendency for local recurrence and may even metastasize to other organ [1, 4, 16]. Radiotherapy and chemotherapy are advocated in cases of partial resection and when surgical specimens show a mesenchymal or myxoid subtype [4, 10, 16, 13, 19]. We didn’t give radiotherapy to our patient since we have achieved a gross total resection and histology concluded to a classical type. Radiosurgery with gamma knife has been also used [1, 13]. Recurrences have been reported [1, 7, 19, 21]; among the 7th reported only one was a classical type. It seems that in the future there will be a place for molecular targets that facilitate tumor invasion and proliferation since recent study found that chronic inflammation in rests of chondrocytes can lead to activation of the P13 kinase pathway and up regulation the $\alpha v\beta 3$ [19].

CONCLUSION

Falcine chondrosarcoma are very rare tumors and can be misdiagnosed with other extra axial tumors. They can have some radiological particularities such as «honeycomb» enhancement. The treatment is mainly based on surgery with a total resection whenever it is possible. Radiotherapy and chemotherapy are advocated in cases with mesenchymal and myxoid histological type and in cases of classical type with partial resection. Prognosis depends on extension of resection and histology.

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